

CLAIMS

Sub B1
5 1. A method for typing a sample of a prion or spongiform encephalopathy disease the method comprising comparing and identifying similar physiochemical properties of the sample with a standard sample of known type.

2. A method as claimed in claim 1 wherein the standard sample of known type is bovine spongiform encephalopathy or Creutzfeldt-Jakob disease.

Sub B2
10 3. A method as claimed in claim 1 or claim 2 wherein the comparison of physiochemical properties comprises a comparison of protease resistance and/or glycoform ratios.

15 4. A method as claimed in any one of the preceding claims wherein the protease resistance is proteinase K resistance.

20 5. A method as claimed in any one of claims 2 to 4 wherein the spongiform encephalopathy is mammalian or chicken derived, in particular, bovine, feline, cervine, ovine, human (or other primate—suitably macaque) or murine derived.

25 6. A method as claimed in any one of claims 1 to 5 wherein the method comprises the steps of subjecting the sample to digestion by a protease, electrophoresing the result of the digestion step and comparing the resulting pattern of the electrophoresis with a standard electrophoresis pattern of a known sample.

7. A method as claimed in any of the preceding claims wherein the typing of the sample comprises a method of diagnosing a disease.

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8. A method as claimed in any one of the preceding claims wherein the sample to be typed is mammalian or chicken derived, in particular derived from a human, (or other primate-suitably macaque) bovine, feline, ovine, cervine, or murine animal.

5 9. A method as claimed in any one of the preceding claims wherein the sample to be typed is derived from brain tissue, other central nervous system tissue, a tissue of the lymphoreticular system (including the spleen, tonsil or lymph node), cerebrospinal fluid and/or the blood.

10 10. A method as claimed in claim 6 wherein the electrophoresis pattern of the known sample has a pattern substantially similar to that of type 4 as shown in figure 4.

15 11. A kit for typing a prion or spongiform encephalopathy sample or diagnosing a prion or spongiform encephalopathy disease, the kit comprising a prion or encephalopathy electrophoresis gel standard and optionally a protease enzyme.

12. A kit as claimed in claim 11 wherein the protease is proteinase K.

20 13. A method for identifying infection in an animal and/or tissue of bovine spongiform encephalopathy the method comprising isolating a prion protein from the animal and/or tissue and identifying that said prion protein can be characterized by having three distinct bands on an electrophoresis gel following proteinase K digestion, the bands comprising i) a band of highest molecular weight in the greatest proportion, ii) a band of lowest molecular weight in the lowest proportion, and iii) a band with a
25 molecular weight between i and ii and of a proportion between i and ii or characterized by having substantially similar glycoform proportions as bovine spongiform encephalopathy.

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14. A method as claimed in claim 13 wherein the animal or tissue is non-bovine.

15. A method as claimed in claim 13 or claim 14 wherein the animal, and/or tissue, from which the prion is sampled is mammalian or chicken derived, in particular, human, (or other primate-suitably macaque) bovine, feline, cervine, ovine, or murine derived."

16. A method as claimed in any one of claims 13 to 15 wherein the prion is derived from brain tissue, other central nervous system tissue, a tissue of the lymphoreticular system (including the spleen, tonsil or lymph node), cerebrospinal fluid and/or the blood.

17. A method for assessing and/or predicting the susceptibility of an animal, in particular a human individual, to bovine spongiform encephalopathy or a derivative thereof, the method comprising the step of determining the genotype of the individual at polymorphic residue 129 of PrP.

18. A method as claimed in claim 17 wherein the determination is whether the individual is homozygous or heterozygous at polymorphic residue 129 of PrP.

19. A method as claimed in claim 17 or claim 18 wherein the determination is whether the individual is homozygous for methionine or valine at polymorphic residue 129 of PrP.

20. A method as claimed in any one of claims 17 to 19 wherein the determination is carried out using DNA obtained from a biological sample.

21. A method as claimed in claim 20 wherein the biological material is blood.

22. A kit for use in assessing and/or predicting the susceptibility of an animal, in particular a human individual, to bovine spongiform encephalopathy or a derivative thereof, which comprises at least one pair of primers suitable for PCR amplification of at least a portion of the gene coding for PrP.

23. A kit as claimed in claim 22, wherein the pair of primers is

5'-GTTTTCCAGTGCCCATCAGTG-3', and

5'-CTATGCACTCATTTCATTATGC-3'

24. A method for typing a sample of a prion or spongiform encephalopathy disease, or for diagnosing a prion or spongiform encephalopathy disease, the method substantially as hereinbefore described with reference to the examples.

25. A kit for typing a prion or spongiform encephalopathy sample or diagnosing a prion or spongiform encephalopathy disease substantially as hereinbefore described with reference to the examples.

26. A method for identifying infection in an animal and/or tissue, as claimed in claim 13, substantially as hereinbefore described with reference to the examples.

27. A method for assessing and/or predicting the susceptibility of an animal, in particular a human, to bovine spongiform encephalopathy or a derivative thereof, substantially as hereinbefore described.

28. A kit for assessing and/or predicting the susceptibility of an animal, in particular a human, to bovine spongiform encephalopathy, substantially as hereinbefore described.

5 29. A method for the prevention or treatment of a prion disease comprising the administration of a compound which inhibits the attachment of sugars to proteins and/or glycoproteins.

10 30. A pharmaceutical agent comprising a compound which inhibits the attachment of sugars to proteins and/or glycoproteins in combination with a pharmaceutically acceptable carrier.

15 31. A compound which inhibits the attachment of sugars to proteins and/or glycoproteins for use as an active pharmaceutical agent.

32. A compound as claimed in claim 31 wherein the pharmaceutical agent is used for the prevention or treatment of a prion disease.

20 33. Use of a compound which inhibits the attachment of sugars to proteins or glycoproteins in the manufacture of a medicament for the prevention or treatment of a prion disease.

25 34. A method as claimed in claim 29, an agent as claimed in claim 30, a compound as claimed in claim 31 or 32 or a use as claimed in claim 33 wherein the compound is deoxynojirimycin or a derivative thereof.

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